

CASE REPORT

A CASE OF RIGHT ATRIAL MYXOMA

WITH SPECIAL REFERENCE TO AN UNUSUAL PHONOCARDIOGRAPHIC FINDING

BY

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There have been 15 cases of right atrial myxoma reported in the English language (Kendall and Symonds, 1952; Bahnson and Newman, 1953; Paquet, 1956; Coates and Drake, 1958; Lyons *et al.*, 1958; Kricilkova *et al.*, 1958; Ellis *et al.*, 1958; Fell, 1958; Hopkins, 1958; Hanlon, 1958; Padhi *et al.*, 1959; Belle, 1959; Ashman *et al.*, 1960; Barham Carter *et al.*, 1960) since Prichard's (1951) review. Surgery was attempted in 12 of these and was successful in 7 (Coates and Drake, 1958; Kricilkova *et al.*, 1958; Ellis *et al.*, 1958; Hopkins, 1958; Hanlon, 1958; Padhi *et al.*, 1959; and Ashman *et al.*, 1960). In this paper a case of myxoma of the right atrium successfully removed under hypothermia is described with an unusual phonocardiographic finding. The differential diagnosis of right atrial myxoma is briefly discussed.

Case Report

A 49-year-old white woman was referred to the Cardiac Clinic of the Johannesburg Hospital on April 24, 1960. For 9 months she had had attacks of sudden pain in the right hypochondrium followed by extreme nausea, breathlessness and a feeling of faintness lasting up to 20 minutes. Breathlessness on exercise had also developed and just before admission she could walk only 50 yards. Her appetite had deteriorated and she had lost 40 lb. in weight. As recently as 3 years previously, her heart had been declared normal by several doctors who had examined her for minor ailments.

On physical examination there was a malar flush with moderate cyanosis of the fingers, toes, lips, and cheeks. The blood pressure was 130/100 mm. Hg, with a mild continuous tachycardia (100 per minute). The jugular venous pressure though not elevated showed a large "a" wave. The heart was not clinically enlarged. On auscultation the first heart sound was very widely split with accentuation of the second (tricuspid) component. At the lower sternal area there was a low-pitched mid-diastolic murmur and an apparent presystolic murmur which extended up to tricuspid valve closure. Both murmurs increased in intensity on inspiration. At the base, both components of the second heart sound were audible but the pulmonary component was soft and was heard only in deep expiration and then with difficulty. The liver was palpable two fingers' breadths below the right costal margin. The central nervous and respiratory systems were normal. On fluoroscopy the pulmonary vasculature was normal, there was no cardiomegaly (cardio-thoracic ratio 48%) but prominence of the right heart border was suggestive of some right atrial dilatation. An electrocardiogram showed a generalized low voltage, a P-R interval of 0.19 seconds and no P wave enlargement. Flattened T waves and sagging S-T segments in leads II, III, AVF, and V4-6 were compatible with myocardial ischaemia.

The clinical picture was thus suggestive of isolated tricuspid stenosis or obstruction which, from the history, was of recent onset and rapidly progressive. An intracardiac neoplasm, causing obstruction at the tricuspid valve orifice, seemed the likely diagnosis.

Special Investigations. Hæmoglobin was 13.4 g. per cent, white cell count 8.300 per c.mm., sedimentation rate (3 tests) 4-15 mm. in 1st hour (Wintrobe). The C-reactive protein was positive (+ + + +) and the urine was normal.

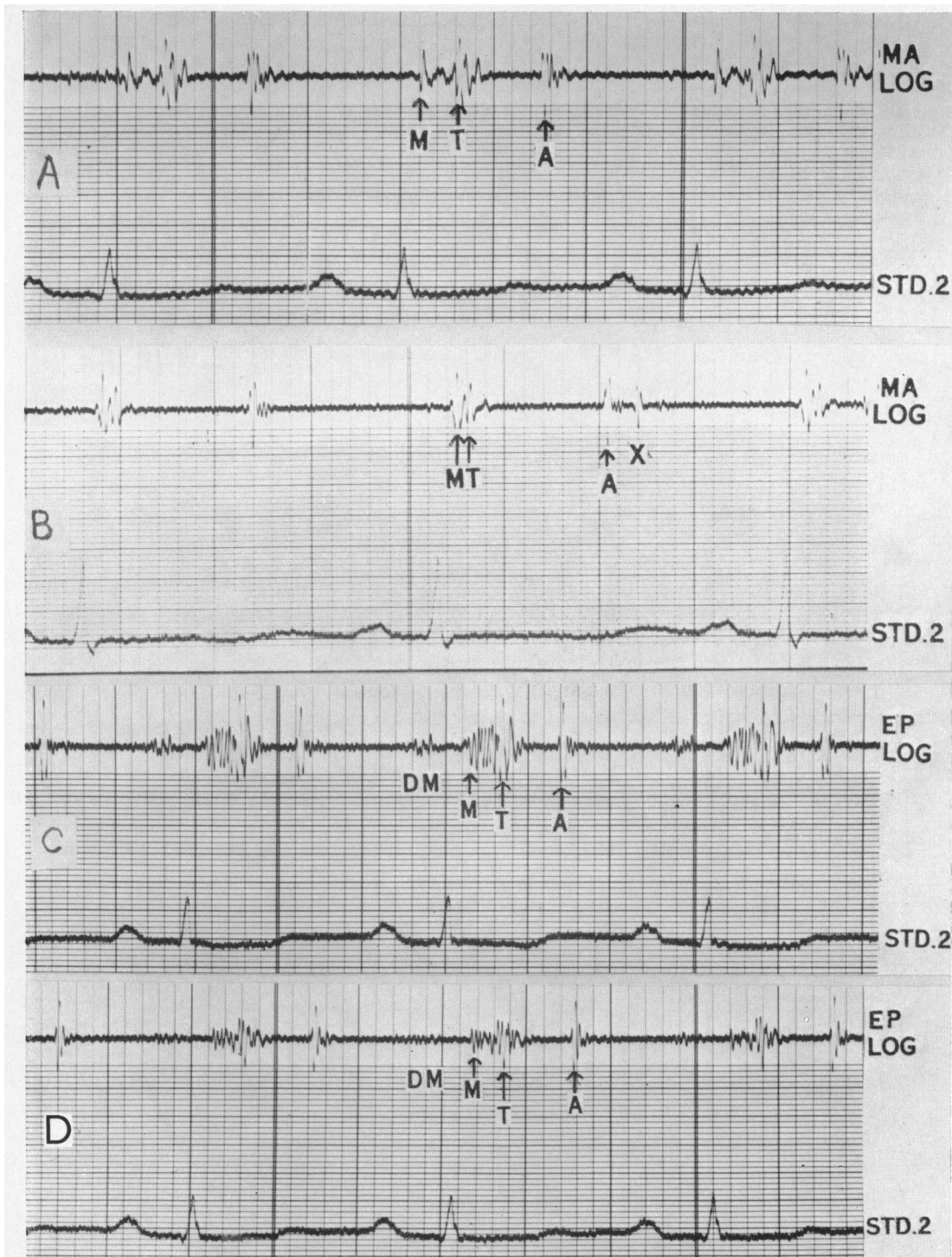


FIG. 1.—Logarithmic phonocardiograms recorded at the mitral area (A) before and (B) after operation. In the preoperative tracing the tricuspid component of the first heart sound is delayed 0.14 sec. after the Q of the simultaneous electrocardiogram and is louder than the mitral component. The postoperative recording shows the relatively soft tricuspid component occurring about 0.02 sec. after the mitral component and about 0.08 sec. after the Q. Preoperative phonocardiogram recorded during (C) deep inspiration and (D) expiration high in the epigastrium. A late diastolic murmur and a “presystolic” murmur, which starts at the time of mitral valve closure and ends at tricuspid valve closure, are shown to increase in intensity with inspiration. For full explanation, see text.

MA=Mitral area. EP=Epigastrium. M=Mitral valve closure. T=Tricuspid valve closure.
A=Aortic valve closure. DM=Late diastolic murmur. X=artefact.

A phonocardiogram (Fig. 1A) confirmed the widely split first sound with the mitral component occurring 0.06 second, and the tricuspid component 0.14 second, after the Q of the simultaneous electrocardiogram. A mid-to-late diastolic murmur and a "presystolic" murmur, which started at about the same time as mitral valve closure and continued up to the delayed tricuspid component, were both maximal on inspiration and were best seen on the recording taken just below the xiphisternum (Fig. 1C, and 1D).

Cardiac catheterization revealed a diastolic gradient of less than 5 mm. Hg between the right atrium and ventricle with a prominent (10 mm. Hg) presystolic wave in the atrial pressure tracing.

Cine-angiocardiology, using a 5 in. image intensifier, demonstrated a large filling defect in the right atrium which moved back and forth with cardiac contraction typical of an atrial myxoma.

Progress

On May 11, 1960, a large angiomyxoma, which was attached by a pedicle to the antero-inferior margin of the coronary sinus, was removed under hypothermia by one of us (D.F.). Convalescence proceeded satisfactorily and the patient was discharged on May 23, 1960. She was last seen in October, 1960, at which time she had no symptoms, had gained 30 lb. in weight, and the physical examination and phonocardiogram (Fig. 1B) were normal.

Discussion

There are certain features common to cases of both right and left atrial myxoma which, when present, are of diagnostic significance:

(1) *Intermittent obstruction* of an atrio-ventricular valve sometimes initiated by postural change results in sudden breathlessness, syncope, convulsions, angina, cyanosis and changing cardiac murmurs (Kirkeby and Leren, 1952; Martin, 1953; Harvey, 1957; Lekisch, 1957; Jackson and Garber, 1958; Towers and Newcombe, 1958; Edwards and Johnson, 1959). Sudden death (Bahnsen *et al.*, 1957; Padhi *et al.*, 1959; Barlow, 1960) and the rare occurrence of cardiac arrhythmias with change in body position (Fawcett and Ward, 1939) probably also depend on atrio-ventricular obstruction by the tumour. The episodes of upper abdominal pain and nausea, which were striking features in the histories of our patient and the case reported by Ashman *et al.* (1960), were again presumably due to acute hepatic distension when the tricuspid orifice was blocked.

(2) A "syndrome" resembling *subacute bacterial endocarditis* is suggested by the common association of anaemia, intermittent pyrexia, raised sedimentation rate and varying cardiac murmurs (Kirkeby and Leren, 1952; Martin, 1953; Lekisch, 1957; MacGregor and Cullen, 1959). The frequent occurrence of systemic emboli (Mills and Philpott, 1951; Goldberg *et al.*, 1952; Steinberg *et al.*, 1953; Chin and Ross, 1957; Gerbode *et al.*, 1958) in left atrial myxoma patients is also compatible with subacute bacterial endocarditis. Pulmonary embolism from a right atrial myxoma (Kendall and Symonds, 1952) seems not to have been encountered so often.

(3) An important *radiological feature* is the relatively mild degree of cardiomegaly in view of the severe clinical state of the patient (Padhi *et al.*, 1959; Ashman *et al.*, 1960). Furthermore, evidence of atrial enlargement is sometimes absent or minimal (Hughes and Faglullah, 1954; Lefcoe *et al.*, 1957; Robertson, 1957). Myxomata seldom calcify but when this does occur the large mass can be seen under the X-ray screen to swing back and forth during the cardiac cycle (Fell, 1958; Hopkins, 1958).

(4) The *electrocardiogram* may show atrial enlargement but this is usually slight and thus incompatible with the apparently severe "mitral or tricuspid stenosis" suggested by the clinical state. No P wave abnormality was seen in some cases of left atrial myxoma (Kirkeby and Leren, 1952; Steinberg *et al.*, 1953; Davis and Andrus, 1954; Cropper and Winstanley, 1955). In several reports of right atrial myxoma, where the electrocardiograms are illustrated (Coates and Drake, 1958; Kricilkova *et al.*, 1958; Belle, 1959; Ashman *et al.*, 1960), the P waves, though somewhat peaked in appearance, do not exceed the normal upper limit of 2.5 mm. (Thomas and Dejong, 1954; Abildskov, 1959).

The Diagnosis of Right Atrial Myxoma

Although Ebstein's anomaly (Coates and Drake, 1958; Belle, 1959) and constrictive pericarditis (Bahnsen and Newman, 1953; Ellis *et al.*, 1958; Padhi *et al.*, 1959) were considered as possible

diagnoses in several previous cases, tricuspid stenosis is the condition which a right atrial myxoma most closely resembles. By far the commonest cause of tricuspid stenosis is, of course, rheumatic heart disease but rheumatic tricuspid stenosis probably never occurs without an associated mitral lesion (Gibson and Wood, 1955) and this in itself should make a diagnosis of isolated tricuspid stenosis suspect (Yater, 1931; Padhi *et al.*, 1959). Congenital tricuspid stenosis is extremely rare (Lewis, 1944) and is very unlikely to be present in patients with a short history of cardiac symptoms such as usually occurs with myxoma. Carcinoid syndrome may cause tricuspid stenosis (Cosh *et al.*, 1959) but the associated pulmonary stenosis and other features will usually serve to distinguish that condition. A familial form of "obstructive cardiomyopathy," recently reported by Hollman *et al.* (1960), has many clinical features suggesting isolated tricuspid stenosis due to the right ventricular inflow tract obstruction. Should such asymmetrical ventricular hypertrophy present without a family history, an angiocardigram would probably be necessary to distinguish it with certainty from a right atrial myxoma.

A 33-year-old man with a presystolic murmur at the sternum maximal on inspiration, low-grade fever, tachycardia, recurrent hæmoptyses, raised sedimentation rate, transient lung shadows and right atrial enlargement radiologically, a normal electrocardiogram and a diastolic gradient at cardiac catheterization of 8 mm. Hg across the tricuspid valve, was believed by Gibson and Wood (1955) to have tricuspid stenosis resulting from disseminated lupus erythematosus. We have been unable to find a similar case reported and the eventual outcome of this patient is unknown (Wood, 1960) but a right atrial myxoma, probably associated with pulmonary emboli, is a possible explanation of that clinical picture. It would seem important that a diagnosis of isolated tricuspid stenosis, whatever the suggested ætiology, should always be queried until an angiogram has excluded an intracardiac neoplasm.

The Phonocardiographic Features

It is now generally agreed that the two major components of the first heart sound are caused by mitral followed by tricuspid valve closure. A most striking feature on the phonocardiogram of our patient was the extremely widely split first heart sound with the tricuspid component occurring 0.08 seconds after mitral valve closure and 0.14 seconds after the Q of the simultaneous electrocardiogram (Fig. 1A). In normal hearts the mitral component of the first heart sound occurs 0.03 to 0.06 seconds after the Q (Leonard *et al.*, 1958; Barlow and Kincaid-Smith, 1960) and Leatham (1954) considers that the tricuspid component occurs 0.02 to 0.03 seconds after mitral valve closure. We are not aware of any reported figures for the time of onset of the tricuspid component in tricuspid stenosis or other conditions which may cause its delay, but in our own experience we have not hitherto observed tricuspid valve closure to occur later than 0.10, or at most 0.11 second after the Q in any condition provided right bundle-branch block was not present.

A presystolic murmur in either mitral or tricuspid stenosis depends on atrial contraction and a consequent flow of blood into the ventricle through the stenosed atrio-ventricular valve. With the onset of ventricular systole and the rise in ventricular pressure, the atrio-ventricular valve closes at the time when ventricular pressure exceeds atrial pressure. Fig. 2, however, shows the simultaneous right atrial pressure, electrocardiogram and phonocardiogram of our patient on which the right ventricular pressure, taken a few seconds earlier, has been superimposed. The tracings reveal that the right atrial pressure is low at the time of the "presystolic" murmur and that the right ventricular pressure exceeds right atrial pressure during this period. This must presumably mean that the "presystolic" murmur is, in fact, due to blood flowing from right ventricle to right atrium. Clearly, in any other circumstances, the tricuspid valve would have closed as soon as ventricular pressure exceeded atrial pressure. In this instance, however, the valve has remained open and allowed blood to regurgitate from ventricle to atrium. It would seem probable that the explanation for this unique situation is that the myxoma protruded through the tricuspid orifice and thus prevented the valve from closing until the point 0.14 seconds after Q. At that time, either due to the high pressure in the right ventricle or by a mechanical swing on its pedicle during cardiac contraction, the myxoma

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